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114.SICKLE CELL DISEASE, SICKLE CELL TRAIT AND OTHER HEMOGLOBINOPATHIES, EXCLUDING THALASSEMIAS: CLINICAL AND EPIDEMIOLOGICAL

A Comprehensive Echocardiographic Portrait of Adult Sickle Cell Disease Patients in the Modern Era

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Background
Cardiovascular complications, especially pulmonary hypertension, are a known cause of mortality in patients with sickle cell disease (SCD). With advances in medicine and the introduction of various disease-modifying medications, the incidence and prevalence of cardiovascular complications are poorly documented.
Aim
To determine rates of cardiovascular complications in patients with SCD in the modern era of disease-modifying medications in a single center.
Method
We performed a retrospective observational single-center study of patients with SCD: all genotyped and ≥16 years old who had a recent echocardiogram performed during the study period of 2019-2023. Demographics, echocardiogram results, and laboratory findings were collected and analyzed. Descriptive statistics are presented in percentages, medians, and interquartile ranges. An unpaired Student's *t*-test and Fisher's exact test were used to determine, where appropriate, the level of significant differences among continuous and categorical variables, respectively, with p value <0.05 considered statistically significant. Data was analyzed using GraphPad Prism version 9.0.2.
Results
As of June 2023, 80 patients (38 male and 42 female, median age 30 years) had undergone a routine transthoracic echocardiogram. Sixty-five patients had the HbSS genotype, 8 HbSC, and 7 HbSB null. Sixty-one patients had received at least one treatment, and 19 (24%) were on concurrent therapy. Hydroxyurea was the common SCD-modifying therapy that the patients received (51 patients), followed by L-glutamine and voxelotor (11 each) and crizanlizumab (9 patients).
In total, 56 (70%) patients had at least one cardiac abnormality. The most common cardiac abnormality was left atrial dilatation (41%), followed by left ventricular hypertrophy (33%), right atrial dilation (24%), and left ventricular dilatation (23%). Pulmonary hypertension was present in 30 (38%) patients with tricuspid regurgitation velocity (TRV) ≥2.5 msec (Table 1).
The cohort of patients with pulmonary hypertension tended to be older (median age 31 years, p=0.0357), have a higher creatinine level (median 0.8, p=0.0414), and have coexisting echocardiographic abnormalities such as left atrial dilatation (70% vs. 24%, p<0.0001) or right atrial dilatation (40% vs. 14%, p=0.0136) than SCD patients without pulmonary hypertension (Table 2).
Conclusion
We present a comprehensive echocardiographic assessment of patients with SCD. Despite advances in the treatment of SCD, there remains a high prevalence of cardiac involvement in SCD patients. We found out that 70% of the patients we reviewed had at least one cardiac abnormality on echocardiography. Left atrial dilatation and left ventricular hypertrophy were the most common cardiac abnormalities noted. Pulmonary hypertension was observed in 37.5% of patients with co-existing right and left atrial enlargement. Larger studies are needed to corroborate these findings.

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Table 1: Demographic and clinical characteristics of the study cohort (n = 80).

Medium age, years (IQR)	30 (24–37)
Sex	
Male	38 (47.5)
Female	42 (52.5)
Genotype	
HbSS	65 (81.3)
HbSC	8 (10.0)
HbS-β thalassemia	7 (8.9)
History	
Hemochromatosis	21 (26.3)
Psychiatric disorders	16 (20.0)
Venous thromboembolism	13 (16.3)
CVA	6 (7.5)
Retinopathy	5 (6.3)
Leg ulcers	3 (3.8)
Treatment	
Received at least 1 treatment	61 (76.3)
Concurrent therapy	19 (23.8)
Hydroxyurea therapy	51 (63.8)
L-glutamate therapy	11 (13.8)
Voxelotor therapy	11 (13.8)
Crizanlizumab therapy	9 (11.3)
Laboratory data	
BNP, pg/mL (IQR)	40 (17–118)
Urine protein/creatinine ratio, mg/g (IQR)	149.5 (87.5–333.7)
Hemoglobin, mg/dL (IQR)	8.8 (7.8–9.98)
LDH, unit/L (IQR)	351 (257.3–465)
Total bilirubin, IU/L (IQR)	2 (1.16–3.8)
Creatinine, mg/dL (IQR)	0.69 (0.54–0.84)
Echocardiographic features	
Left ventricle ejection fraction, % (IQR)	60 (55–61.7)
TRV, m/sec (IQR)	2.4 (2.12–2.7)
Right ventricular systolic pressure (RVSP), mmHg (IQR)	28 (22–33.25)
Left atrial dilatation	33 (41.3)
Left ventricle dilatation	18 (22.5)
Left ventricle hypertrophy	26 (32.5)
Right atrial dilatation	19 (23.8)
Pulmonary hypertension with TRV >2.5	30 (37.5)
At least one echocardiographic abnormality	56 (70)

Note. Data are no. (%) unless otherwise indicated.4

Table 2: Patients with versus without pulmonary hypertension.

	TRV ≥2.5, n=30	TRV <2.5, n=50	p-value
Medium age, years (IQR)	31 (24.8–44.8)	29 (23.8–34.3)	0.0357
Sex			
Male	15 (50)	23 (46)	0.8185
Female	15 (50)	27 54)	
Genotype			
HbSS	23 (77)	42 (84)	0.5552
HbS-β thalassemia	4 (13)	3 (6)	
HbSC	3 (10)	5 (10)	
History			
Hemochromatosis	10 (33)	11 (22)	0.3011
Psychiatry disorders	3 (10)	13 (26)	0.1471
Venous thromboembolism	7 (23)	6 (12)	0.2188
CVA	1 (3)	5 (10)	0.4018
Retinopathy	4 (13)	1 (2)	0.0629
Leg ulcers	2 (7)	1 (2)	0.5527
Treatment			
Received at least 1 treatment	24 (80)	37 (74)	0.5982
Concurrent therapy	8 (27)	11 (22)	0.7868
Hydroxyurea therapy	20 (67)	31 (62)	0.8110
L-glutamate therapy	2 (7)	7 (14)	0.4714
Voxelator therapy	5 (16)	6 (12)	0.7387
Crizanlizumab therapy	6 (20)	5 (10)	0.3146
Laboratory data			
BNP, pg/mL (IQR)	59 (22.3–173)	29 (16–106.5)	0.3356
Urine protein/creatinine ratio, mg/g (IQR)	129 (98.8–50.4)	156.5 (83.3–256.5)	0.3429
Hemoglobin, mg/dL (IQR)	8.4 (7.4–9.9)	9.1 (7.9–10.1)	0.3305
LDH, unit/L (IQR)	357.5 (265.6–464.3)	339 (253.5–479.3)	0.8987
Total bilirubin, IU/L (IQR)	1.8 (0.88–3.35)	2.1 (1.4–3.4)	0.9496
Creatinine, mg/dL (IQR)	0.8 (0.57–0.90)	0.6 (0.5–0.8)	0.0414
Echocardiographic features			
Left ventricle ejection fraction, % (IQR)	59 (55–60.5)	60 (55–61.9)	0.2248
PASP/RVSP, mmHg (IQR)	34.5 (29–40)	22 (17.4-26)	<0.0001
Left atrial dilatation	21 (70)	12 (24)	<0.0001
Left ventricle dilatation	10 (33)	8 (16)	0.0981
Left ventricle hypertrophy	11 (37)	15 (30)	0.6243
Right atrial dilatation	12 (40)	7 (14)	0.0136

Note. Data are no. (%) unless otherwise indicated.

Figure 1

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